Outcomes of patients born with single-ventricle physiology and aortic arch obstruction: The 26-year Melbourne experience

Melissa G. Y. Lee, BMedSc,[a] Christian P. Brizard, MD,[a] John C. Galati, PhD,[b] Ajay J. Iyengar, MBBS, BMedSc,[a] Sandeep S. Rakhra, MBBS, BMedSc,[a] Igor E. Konstantinov, MD, PhD,[a] Andreas Pflaumer, MD,[c] and Yves d'Udekem, MD, PhD[a]

Background: To review the long-term outcomes of patients born with single-ventricle physiology and aortic arch obstruction.

Methods: Follow-up of 70 consecutive neonates undergoing single-ventricle palliation and arch repair, excluding hypoplastic left heart syndrome, between 1983 and 2008, was reviewed. Dominant arch anomalies were coarctation (n = 48), interrupted arch (n = 10), and hypoplastic arch alone (n = 12). Neonatal Damus procedure with arch repair and shunt became the dominant approach, being performed in 1 (10%) of 10 in 1983 to 1989, 9 (32%) of 28 in 1990 to 1999, and 23 (72%) of 32 in 2000 to 2008.

Results: All patients underwent an initial procedure at a median of 6 days (range, 4-12 days): pulmonary artery banding and arch repair (n = 35); Damus, arch repair, and shunt (n = 33); and other (n = 2). Twenty-six patients died before Fontan completion. Of the 34 survivors of initial banding, 17 (50%) later required a Damus and 4 (12%) required subaortic stenosis relief. Forty patients underwent Fontan completion at a median age of 5 years (range, 4-7 years). After a mean of 5 ± 6 years after Fontan, there was 1 hospital death and 1 Fontan takedown. Overall survival was similar if patients initially underwent a Damus or pulmonary artery banding ($P = .3$). Overall survival at 10 years was 53% (95% confidence interval, 42%-67%).

Conclusions: Patients born with single-ventricle physiology and arch obstruction have a high risk of mortality in the first years of life. Their outcomes seem excellent once they reach Fontan status. It is likely that, in patients with single-ventricle and arch obstruction, strategies to avoid systemic outflow tract obstruction should be implemented in early life, and regular monitoring of blood pressure is warranted. (J Thorac Cardiovasc Surg 2014;148:194-201)

CHD

It is widely accepted that aortic arch obstruction is associated with poor outcomes in patients with single-ventricle physiology because of the potential for myocardial hypertrophy and subaortic stenosis in the first year of life.[1,2] There has been much debate on the best approach for these patients in the neonatal period. All teams agree that the aortic arch obstruction should undergo an early relief.[3] There is, however, contention between those who favor the use of an extensive Norwood-type operation, including an aortic arch repair, a Damus-Kaye-Stansel (DKS) procedure, and a systemic-pulmonary shunt and those who prefer to perform an initial banding of the pulmonary artery (PA) at the time of the arch repair and reserve later completion of a DKS procedure to those developing subaortic obstruction. The presence of coarctation or arch obstruction should strongly suggest the potential for systemic obstruction at the bulboventricular foramen,[4] and previous studies have demonstrated that most patients who undergo initial PA banding will ultimately develop subaortic stenosis early in life, necessitating further intervention.[5-8] Yet, initial banding is still favored by many teams today.[6-8] An arterial switch operation may be performed in a few of these patients, but they have a high propensity to develop subaortic stenosis after this operation, and the adjustment of pulmonary blood flow is unpredictable.[9,10]

We have realized that up to half of the patients with biventricular circulation undergoing aortic arch repair may require reintervention in the decades after the initial intervention.[11-13] We know that the development of diastolic dysfunction consequential to the arch obstruction is a factor leading to the failure of the Fontan circulation, and we wondered whether the patients requiring aortic arch

From the Departments of Cardiac Surgery[a] and Cardiology,[c] The Royal Children's Hospital; the Department of Paediatrics, University of Melbourne; and the Murdoch Childrens Research Institute, Melbourne, Australia; and the Clinical Epidemiology and Biostatistics Unit,[b] Murdoch Childrens Research Institute; and the Department of Mathematics and Statistics, La Trobe University, Melbourne, Australia.

This study was supported by the Victorian Government's Operational Infrastructure Support Program. Dr d'Udekem is a Career Development Fellow of The National Heart Foundation of Australia (CR 10M 5339). Dr Iyengar is a cofounded Postgraduate Scholar of the National Health and Medical Research Council/National Heart Foundation of Australia (APP1038802), and holds an Royal Australian College of Surgeons Catherine Marie Enright Kelly Postgraduate Scholarship.

Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication Feb 5, 2013; revisions received June 4, 2013; accepted for publication July 26, 2013; available ahead of print Sept 26, 2013.

Address for reprints: Yves d'Udekem, MD, PhD, Department of Cardiac Surgery, The Royal Children's Hospital, Flemington Rd, Parkville, Melbourne 3052, Victoria, Australia (E-mail: yves.dudekem@rch.org.au).

0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2013.07.076

Abbreviations and Acronyms

BCPS = bidirectional cavopulmonary shunt
BT = Blalock-Taussig
CI = confidence interval
DKS = Damus-Kaye-Stansel
DORV = double-outlet right ventricle
HR = hazard ratio
PA = pulmonary artery

surgery who undergo single-ventricle palliation would have poor outcomes.[14] Little is known of the progression of these patients toward the Fontan pathway, and there has been no information about their outcome beyond Fontan completion.

This study reviews our experience of patients born with single-ventricle physiology and neonatal aortic arch surgery during a period of 26 years, during which time we gradually adopted Norwood-type procedures as our primary approach.

METHODS

Study Population

The design of the study was approved by The Royal Children's Hospital Human Research and Ethics Committee, and the need for consent was waived because of the retrospective nature of the study.

The hospital database was screened to identify the patients who underwent single-ventricle palliation and aortic arch repair for coarctation, interrupted aortic arch, or aortic arch hypoplasia at The Royal Children's Hospital between 1983 and 2008. Patients with hypoplastic left heart syndrome were excluded because of the severity of this condition and associated poor early outcomes in the first era of the study. Seventy patients were identified: 46 (66%) were male and 24 (34%) were female.

Morphologic Data

Morphologic diagnoses of the patients were established by 2-dimensional echocardiography and Doppler flow imaging, and confirmed on macroscopic visualization at the initial neonatal palliation. Characteristics of the patients are displayed in Table 1.

Aortic coarctation was defined as a discrete narrowing at the isthmus of the arch between the left subclavian artery proximally and the ductus arteriosus distally. *Interrupted aortic arch* was defined as an anatomic lack of continuity between the proximal and distal segments of the aortic arch. The arch was considered hypoplastic if the echocardiographic report, the cardiologist's report, or the surgical notes labeled it as hypoplastic, or if any portion of the arch had a z score diameter of less than −2.0.

Surgical Management

All patients underwent palliation in the neonatal period at a median age of 6 days (range, 4-12 days), with the goal to ultimately achieve Fontan circulation: 33 (47%) had a DKS procedure; 35 (50%) had a PA band and aortic arch repair, with 4 of them undergoing the arch repair as a previous separate procedure; and 2 (3%) had arterial switch and aortic arch repair.

A strategy of performing a neonatal DKS procedure became the dominant approach during the study period, being performed in 1 (10%) of 10 patients in the initial years of 1983 to 1989, in 9 (32%) of 28 patients in 1990 to 1999, and in 23 (72%) of 32 patients in the recent 2000 to 2008 period (Figure 1). Consequently, PA banding has become less favored in recent times, being performed in 8 (80%) of 10 patients in 1983 to 1989, 18 (64%) of 28 patients in 1990 to 1999, and in only 9 (28%) of 32 patients in 2000 to 2008.

The DKS procedure involved complete transection of both the PA and the aorta, followed by the anastomosis of these 2 vessels such that no more than one third of the smaller of the 2 vessels was sutured to the larger vessel. The descending aorta was anastomosed to the ascending aorta in an end-to-side manner in most of the cases. Additional pericardial patch was added to the arch repair in 21 patients (64%) using homograft pericardium in 14 and autologous pericardium in 7.

Follow-up

The files and echocardiographic reports of the patients were reviewed, and their follow-up was gathered from hospital databases and their referring cardiologists.

Hospital mortality was defined as death that occurred within 30 days after surgery or during hospital stay. *Late mortality* was defined as death that occurred after this period.

Resting hypertension for children and adolescents was defined as a systolic or diastolic blood pressure of greater than the 95th percentile for age and height, and *prehypertension* was defined as between the 90th and 95th percentile or if blood pressure was greater than 120/80 mm Hg.[15] *In adults, resting hypertension* was defined as a systolic blood pressure of greater than 140 mm Hg or a diastolic blood pressure of greater than 90 mm Hg; and *prehypertension* was defined as a systolic blood pressure between 120 and 139 mm Hg or a diastolic blood pressure between 80 and 89 mm Hg.[16]

Aortic arch reobstruction was defined as a peak gradient exceeding 25 mm Hg across the repair site on echocardiogram, or an upper limb to lower limb blood pressure gradient of more than 20 mm Hg.

Statistical Analysis

All data were exported to and analyzed using STATA, version 12.1 (Stata Corporation, College Station, Tex). Data quoted in the text are summarized as either mean ± SD or median (interquartile range). A nonpaired Student *t* test, a Pearson χ^2 test (when appropriate), or a Fisher exact test was used to compare patients who underwent neonatal DKS with patients who underwent PA banding for each of the characteristics listed in Table 1. A univariable analysis of risk factors for hospital mortality after initial surgery was assessed using either a Fisher exact test or logistic regression. Cox regression analysis was used to examine the association of reintervention for systemic outflow tract obstruction and mortality with all collected patient and surgical characteristics. Because of the few outcomes, it was not feasible to perform multivariable risk analyses.

RESULTS

The postoperative courses of the 70 patients in this study are summarized in Figure 2. The mean follow-up from initial neonatal palliation was 7 ± 7 years.

Initial Neonatal Surgery

Comparative characteristics of the 70 patients at their first palliative procedure are given in Table 1. The sources of pulmonary blood flow used for the DKS procedure were as follows: modified Blalock-Taussig (BT) shunt (n = 30) and right ventricle–pulmonary artery shunt (n = 3).

Fifteen patients (21%) had the following 16 concomitant procedures at their initial neonatal surgery: atrial septectomy (n = 9), cor triatriatum repair (n = 2), subaortic stenosis relief (n = 2), tricuspid valve repair (n = 1),

CHD

TABLE 1. Patient characteristics

Characteristics	Neonatal DKS (n = 33)	Neonatal PAB + AR (n = 35)	Neonatal ASO + AR (n = 2)	*P* value DKS/PAB
Demographics				
Sex				
Male	22 (67)	23 (66)	1 (50)	.9
Female	11 (33)	12 (34)	1 (50)	
Birth weight, kg	3.3 ± 0.6 (n = 24)	3.1 ± 0.8 (n = 17)	3.3 (n = 1)	.5
Age at surgery, d	16 ± 40	19 ± 53	51 ± 54	.8
Dominant diagnosis				
Tricuspid atresia	12 (36)	4 (11)	0 (0)	.02
Double-inlet left ventricle	10 (30)	16 (46)	2 (100)	.2
Double-outlet right ventricle	2 (6)	4 (11)	0 (0)	.4
AVSD	3 (9)	4 (11)	0 (0)	.8
MS/MA + VSD	3 (9)	1 (3)	0 (0)	.3
TGA + hypoplastic RV	2 (6)	1 (3)	0 (0)	.5
Aortic stenosis	1 (3)	0 (0)	0 (0)	.3
Other	0 (0)	5 (15)	0 (0)	.02
Aortic arch anomaly				
HAA only	11 (33)	1 (3)	0 (0)	.001
Coarctation ± HAA	14 (42)	31 (89)	2 (100)	.0001
IAA ± HAA	8 (25)	3 (8)	0 (0)	.2
Type A	4	1	0	
Type B	4	2	0	
Any HAA	27 (82)	16 (46)	2 (100)	.002
Cardiac anomalies				
Situs				
Solitus	32 (97)	32 (91)	2 (100)	.3
Inversus/ambigous	1 (3)	3 (9)	0 (0)	
Isomerism	1 (3)	4 (13)	0 (0)	.2
Ventricular type				
Left	21 (63)	16 (46)	1 (50)	.1
Right	12 (36)	14 (40)	1 (50)	.8
Biventricular	0 (0)	5 (14)	0 (0)	.02
TGA	26 (79)	19 (61)	2 (100)	.2
Bilateral superior vena cava	4 (67)	4 (66)	0 (0)	.6

Data are given as number (percentage) or mean ± SD. *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding; *AR*, arch repair; *ASO*, arterial switch operation; *AVSD*, atrioventricular septal defect; *MS*, mitral stenosis; *MA*, mitral atresia; *TGA*, transposition of the great arteries; *RV*, right ventricle; *HAA*, hypoplastic aortic arch; *IAA*, interrupted aortic arch, *VSD*, Ventricular septal defect.

atrioventricular septal defect repair (n = 1), and anomalous left coronary artery from the PA repair (n = 1). Eight of the patients were undergoing PA banding.

Nine patients (13%) died within the first 30 days or during hospital stay. Causes of death were low cardiac output syndrome in 8 and septicemia in 1. The early mortality rate of patients treated with neonatal DKS compared with patients treated with the PA banding procedure was 24% (8/33) versus 3% (1/35), respectively ($P = .01$). Other risk factors for early mortality were having a dominant right ventricle ($P < .001$), with all 9 cases of mortality having dominant right ventricle physiologic features and a double-outlet right ventricle (DORV) ($P = .03$).

Of the 61 survivors, 8 (13%) required 11 cardiac procedures while awaiting a superior cavopulmonary anastomosis operation, and 7 of them had undergone neonatal PA banding. Procedures included subaortic stenosis relief (n = 2), right modified BT shunt insertion (n = 2), resection of aneurysm (n = 1), atrial septectomy (n = 1), arterial switch operation (n = 1), PA band tightening (n = 1), mitral valve replacement (n = 1), PA reconstruction (n = 1), and pacemaker insertion (n = 1).

Of the 61 survivors of the initial neonatal surgery, 4 (7%) died in the interim while awaiting a superior cavopulmonary anastomosis procedure; 2 had undergone neonatal DKS, and 2 had PA banding. The cause of death in 3 patients was undetermined, and 1 patient died of heart failure from severe atrioventricular regurgitation.

Superior Cavopulmonary Anastomosis

There were 57 survivors of an initial neonatal procedure (81%) reaching a second palliation for superior cavopulmonary anastomosis. In the 1980s, 5 of the survivors who had undergone initial PA banding underwent a

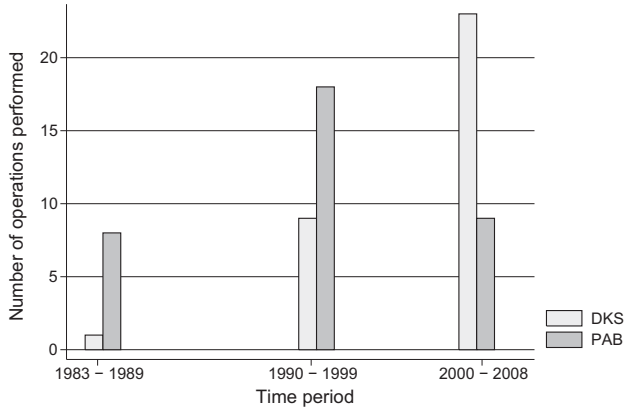

FIGURE 1. Type of initial neonatal palliation performed by era. *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding.

Fontan procedure without prior superior cavopulmonary anastomosis. Consequently, 52 patients underwent superior cavopulmonary anastomosis at a median age of 9 months.[5-18] The median age at superior cavopulmonary anastomosis was 16 months[8-29] for those who had undergone initial PA banding. The type of superior cavopulmonary anastomosis operation performed is summarized in Table 2.

Of the 52 patients, 30 (58%) underwent 33 concomitant procedures: DKS (n = 12), PA reconstruction (n = 6), atrial septectomy (n = 5), aortic arch reintervention (n = 4), subaortic stenosis relief (n = 1), mitral valve repair (n = 1), tricuspid valve repair (n = 1), DORV repair (n = 1), right modified BT shunt insertion (n = 1), and pacemaker insertion (n =1). After DKS, no patient developed significant regurgitation of his or her semilunar valves.

Of the 52 patients, 4 (8%) who underwent a superior cavopulmonary anastomosis procedure died within the first 30 days or during hospital stay. One patient who required a redo left bidirectional cavopulmonary shunt (BCPS) and common atrioventricular valve repair within the same hospital admission for BCPS died subsequently of heart failure with atrioventricular valve regurgitation. The remaining cause of death was low cardiac output.

The hospital mortality after superior cavopulmonary anastomosis was 4% (1/23) for patients treated with neonatal DKS compared with 11% (3/27) for patients treated with the PA banding procedure ($P = .6$).

Three patients who required a superior cavopulmonary anastomosis takedown a mean of 11 ± 17 months after the superior cavopulmonary anastomosis operation subsequently died. Two of these patients had undergone neonatal PA banding.

Of the 48 superior cavopulmonary anastomosis survivors, 7 (3 with initial PA banding) required 7 cardiac procedures while awaiting Fontan completion: atrial septectomy (n = 1), aortic arch repair for recoarctation (n = 1), pacemaker insertion (n = 1), reconstruction of subclavian and

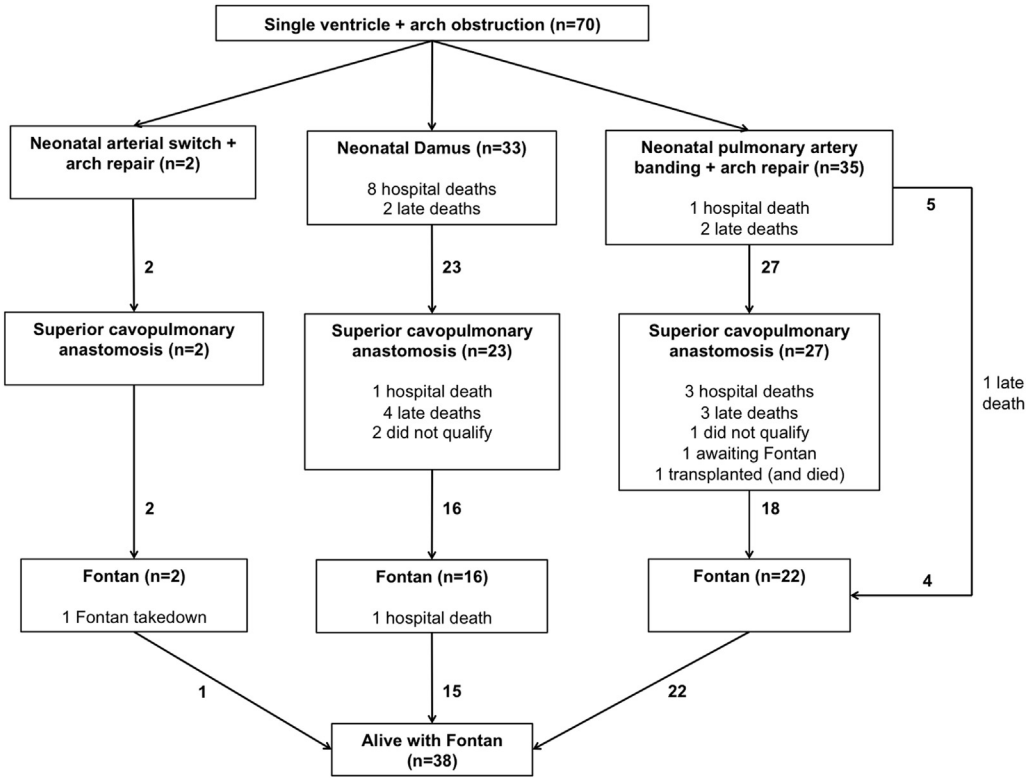

FIGURE 2. Flowchart of patients' interventions and outcomes.

TABLE 2. Superior cavopulmonary anastomosis procedures

	Neonatal DKS (n = 23)	Neonatal PAB + AR (n = 27)	Neonatal ASO + AR (n = 2)
Procedure			
BCPS	20 (87)	24 (89)	2 (100)
Bilateral BCPS	3 (13)	0 (0)	0 (0)
Classic Glenn	0 (0)	2 (7)	0 (0)
Kawashima	0 (0)	1 (4)	0 (0)
Concomitant DKS	0 (0)	15 (58)	0 (0)

Data are given as number (percentage). *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding; *AR*, arch repair; *ASO*, arterial switch operation; *BCPS*, bilateral cavopulmonary shunt.

internal jugular veins (n = 1), mitral valve repair (n = 1), PA reconstruction (n = 1), and right modified BT shunt insertion (n = 1).

In addition, 2 of the 5 survivors of initial palliation who did not undergo a superior cavopulmonary anastomosis operation required 4 cardiac procedures while awaiting Fontan completion: DKS (n = 2), atrial septectomy (n = 1), and PA reconstruction (n = 1).

Of the 48 survivors, 7 (15%) died in the interim while awaiting Fontan completion, including the 3 patients who required a superior cavopulmonary anastomosis takedown and 1 patient who was denied Fontan completion and required heart transplantation. Of the 7 mortalities, 3 (43%) had undergone neonatal PA banding, and 3 (43%) had undergone an additional cardiac procedure.

In addition, 1 of the 5 survivors of initial palliation who did not undergo a superior cavopulmonary anastomosis operation died of an undetermined cause while awaiting Fontan completion. This patient had undergone an additional cardiac procedure.

Fontan Operation

Four patients with PA banding proceeded directly to Fontan completion after the initial neonatal operation. Of the 70 patients in this study, 40 (57%) underwent Fontan completion a mean of 4 ± 1 years after superior cavopulmonary anastomosis at a median age of 5 years.[4-6] Eleven patients (7 with initial PA banding) underwent 12 concomitant procedures at Fontan completion: atrial septectomy (n = 3), PA reconstruction (n = 3), tricuspid valve repair (n = 2), subaortic stenosis relief (n = 1), DKS (n = 1), mitral valve repair (n = 1), and pacemaker insertion (n = 1).

One patient died 3 days after hospital discharge, with no known cause. One patient required a Fontan takedown to BCPS and thrombectomy of the left PA 14 days after the operation.

Follow-up

Four Australian patients and one international patient were lost to follow-up. Of the 38 survivors with intact Fontan circulation, 33 (87%) were followed up for a mean of 10 ± 6 years after initial neonatal surgery and 5 ± 6 years after Fontan completion. The median age at last follow-up was 9 years.[6-17]

There were no late deaths. All 33 observed patients were in New York Heart Association class I or II. Of the 33 observed patients, 5 (15%) developed an arrhythmia a mean of 6 ± 7 years after Fontan completion and 3 required pacemaker insertion (all 3 patients had neonatal PA banding). Four patients required 4 other cardiac procedures after Fontan completion: right pleurodesis (n = 1), aortic arch reconstruction (n = 1), DKS (n = 1), and left BT shunt insertion for ongoing cyanosis (n = 1).

Hypertension

Resting blood pressure measurements at last follow-up were available in 20 patients (61%). Ten patients (50%) had abnormal blood pressure measurements: 5 had prehypertension, and 5 had hypertension. All 5 patients with hypertension and 3 with prehypertension had undergone neonatal PA banding. The proportion of patients with abnormal blood pressure among patients treated with neonatal DKS compared with patients treated with the PA banding procedure was 25% (2/8) versus 67% (8/12), respectively ($P = .17$).

Reobstruction

Measurements of the maximum velocity in the descending aorta on echocardiogram at last follow-up were available in only 15 patients (45%). One patient who had initial PA banding developed arch reobstruction on echocardiography and was also hypertensive.

Another patient required an aortic arch reconstruction with a Dacron patch for reobstruction on echocardiogram 8 years after Fontan. This patient had undergone neonatal PA banding and was prehypertensive on last follow-up, despite a normal descending aorta velocity on echocardiogram.

Overall Systemic Outflow Tract Obstruction

Of the 70 patients, 21 (30%) in this study required 22 additional procedures to relieve a systemic outflow tract obstruction: DKS shunt after neonatal palliation (n = 17) and subaortic stenosis relief (n = 5). All 21 patients had undergone neonatal PA banding ($P < .001$), representing 62% of the 34 hospital survivors of PA banding.

Of the 21 patients, 16 (76%) underwent Fontan completion and 1 was awaiting Fontan completion. The overall mortality rate for patients who required systemic outflow tract relief was 24% (5/21); 4 died before Fontan completion, and 1 died after Fontan completion.

Overall freedom from systemic outflow tract reintervention at 1, 5, and 10 years was 79% (95% confidence interval [CI], 66%-88%), 66% (95% CI, 52%-78%), and 55%

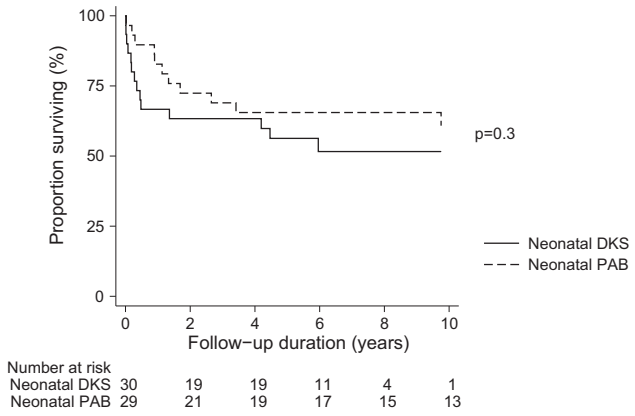

FIGURE 3. Kaplan-Meier survival curve. *DKS*, Damus-Kaye-Stansel; *PAB*, pulmonary artery banding.

(95% CI, 36%-71%), respectively. Risk factors for overall systemic outflow tract reintervention on univariate analysis other than neonatal PA banding were having double-inlet left ventricle (hazard ratio [HR], 2.9; 95% CI, 1.6-7.6; $P = .03$) and having coarctation (HR, 9.0; 95% CI, 1.2-67.5; $P = .03$).

Overall Survival

Overall survival at 1, 5, and 10 years was 77% (95% CI, 62%-83%), 60% (95% CI, 48%-72%), and 53% (95% CI, 42%-67%), respectively. Survival at 1, 5, and 10 years for patients who underwent neonatal DKS and patients who had neonatal PA banding was 63% (95% CI, 48%-78%) versus 82% (95% CI, 65%-92%), 55% (95% CI, 38%-72%) versus 63% (95% CI, 47%-78%), and 52% (95% CI, 32%-68%) versus 62% (95% CI, 42%-77%), respectively (Figure 3). There was no significant difference in overall mortality between patients who underwent neonatal DKS and patients who had neonatal PA banding ($P = .3$). Risk factors for overall mortality on univariate analysis were DORV (HR, 3.4; 95% CI, 1.3-9.1; $P = .02$), atrioventricular septal defect (HR, 3.8; 95% CI, 1.5-9.6; $P = .005$), dominant right ventricle (HR, 3.1; 95% CI, 1.4-6.9; $P = .006$), isomerism (HR, 3.1; 95% CI, 1.1-8.9; $P = .04$), transposition of the great arteries (HR, 0.31; 95% CI, 0.15-0.66; $P = .003$), superior cavopulmonary anastomosis takedown (HR, 6.1; 95% CI, 1.9-19.8; $P = .003$), and ever having an atrial septectomy procedure (HR, 0.22; 95% CI, 0.05-0.94; $P = .04$).

DISCUSSION

There is substantial evidence that the rate of reobstruction of arches repaired in infancy is considerable and that many of these patients will develop hypertension, even if they do not have arch reobstruction.[11,13,17] It is likely that the development of arch obstruction and hypertension will more profoundly affect patients with single-ventricle physiologic features than patients with biventricular circulation, because it will undoubtedly generate systemic ventricular hypertrophy and increased diastolic dysfunction, which will directly affect the passive flow of the blood through the lungs and cause any type of cavopulmonary circulation to fail.[14] We reviewed the long-term outcomes of patients who had undergone single-ventricle palliation and aortic arch repair to find out to what extent aortic arch obstruction would adversely affect these outcomes, especially in terms of progression to the Fontan pathway and Fontan failure.

This is a historic review and, as such, was not designed to compare the different approaches offered to neonates presenting with aortic arch obstruction and single-ventricle physiologic features. However, there are such striking findings in the early outcomes of this population that it is impossible not to debate on the contentious topic of the best initial approach to offer these patients. In the 1980s, it already became clear that up to 85% of patients born with single-ventricle physiologic features and arch obstruction, who would, by definition, have high pulmonary blood flow necessitating banding of their PA, would develop subaortic obstruction. And, it was already realized at the time how adversely it would compromise their future Fontan circulation.[5] Therefore, in the 1990s, some teams advocated the use of more radical Norwood-type operations to alleviate the subsequent development of subaortic stenosis and avoid its associated ventricular thickening.[18-20] Some excellent results were reported, yet today there is still no agreement between teams favoring this latter Norwood-type approach and those who still favor arch repair and banding in the neonatal period and await the development of subaortic obstruction to proceed to a DKS connection. Mortality after initial DKS remained high, with most series reporting early mortality of 19% to 38%.[18,21,22]

Our experience confirms that performing an initial PA banding is an approach with less initial mortality than the approach consisting of a Norwood-type procedure: the arch repair, DKS, and pulmonary artery shunting. It also confirms that this PA band approach may expose the patients to higher rates of late mortality and late complications. The same proportion of patients ultimately died after both approaches, but the mortality occurred within the first hospital stay after initial DKS, whereas it occurred later, after first hospital discharge, after the PA banding approach. Our reported experience spans more than 3 decades, during which time patient selection and perioperative management varied enormously. During the course of these 3 decades, we have switched from a strategy of initial PA banding to neonatal Norwood-type procedures. We have demonstrated that, in the past decade, our survival after Norwood surgery dramatically improved.[23] We, therefore, believe that this retrospective study spanning more than

3 decades, which includes our learning curve with the Norwood-type procedures, does not allow comparison between both approaches. We were favorably impressed by the good survival of the patients undergoing initial PA banding in the early 1980s, and we may favor this approach in selected cases. Some may be influenced by our results to favor initial PA banding, knowing that this strategy will expose patients to a higher risk of later complications. It has been repeatedly demonstrated that survival after Norwood surgery is related to surgeon and center volume of cases, and some may want to avoid the risks of this strategy.[23-26] Patients undergoing this strategy should undergo an early cavopulmonary connection with near-systematic use of a DKS connection at BCPS. Some may also favor early arterial switch, which has been shown in some case series to give favorable results.[27] Our opinion is still nonetheless that performing a DKS will prevent the development of subaortic obstruction and that shunting will better control pulmonary blood flow than PA banding; we will, therefore, pursue our approach of Norwood type in most of these patients.

The higher proportion of patients with hypertension in the group of patients undergoing initial banding is likely the consequence of a less radical relief of the arch obstruction in this group. In addition, 21 of the 34 hospital survivors of an initial banding (62%) required a relief of subaortic obstruction. It is, therefore, likely that suboptimal relief of the arch obstruction and delayed relief of the subaortic obstruction will generate deleterious ventricular hypertrophy and affect the risk of failure of the Fontan circulation, even though we cannot substantiate this hypothesis at this stage. In addition, it has been demonstrated that the DKS procedure can still be effectively performed after banding,[28,29] but it is undeniable that banding may compromise the quality of these valves.

It was surprising for us to observe how poorly our patients were followed up for the development of aortic arch obstruction and hypertension. At this stage, we could not yet demonstrate any deleterious outcomes after Fontan surgery, which we believe is related to an aggressive policy toward relief of arch obstruction and subaortic stenosis. Nonetheless, it is striking that these patients are still at risk of hypertension. Specific follow-up focused on aortic arch obstruction, and the development of hypertension should be mandatory in patients born with single ventricles and aortic arch obstruction.

Limitations

This series describes operations performed over a long period. Despite being the largest report of such a patient population, this report cannot eliminate patient selection bias and is, therefore, not the ideal substrate to compare different surgical approaches. Conclusions drawn from the occurrence of late hypertension in this patient population would be stronger if many of these patients had a systematic screening of their blood pressure using 24-hour ambulatory blood pressure monitoring.[11,17]

In conclusion, patients born with single-ventricle physiologic and arch obstruction have a high risk of mortality in the first years of life. Their outcomes seem excellent once they reach Fontan status. It is likely that, in patients with single-ventricle and arch obstruction, strategies to avoid systemic outflow tract obstruction should be implemented in early life, and regular monitoring of blood pressure is warranted.

References

1. Caspi J, Coles JG, Rabinovich M, Cohen D, Trusler GA, Williams WG, et al. Morphological findings contributing to a failed Fontan procedure: twelve-year experience. *Circulation*. 1990;82(suppl):IV177-82.
2. Gentles TL, Mayer JE Jr, Gauvreau K, Newburger JW, Lock JE, Kupferschmid JP, et al. Fontan operation in five hundred consecutive patients: factors influencing early and late outcome. *J Thorac Cardiovasc Surg*. 1997; 114:376-91.
3. Cohen M, Fuster V, Steele PM, Driscoll D, McGoon DC. Coarctation of the aorta: long-term follow-up and prediction of outcome after surgical correction. *Circulation*. 1989;80:840-5.
4. Matitiau A, Geva T, Colan SD, Sluysmans T, Parness IA, Spevak PJ, et al. Bulboventricular foramen size in infants with double-inlet left ventricle or tricuspid atresia with transposed great arteries: influence on initial palliative operation and rate of growth. *J Am Coll Cardiol*. 1992;19:142-8.
5. Freedom RM, Benson LN, Smallhorn JF, Williams WG, Trusler GA, Rowe RD. Subaortic stenosis, the univentricular heart, and banding of the pulmonary artery: an analysis of the courses of 43 patients with univentricular heart palliated by pulmonary artery banding. *Circulation*. 1986;73:758-64.
6. Webber SA, LeBlanc JG, Keeton BR, Salmon AP, Sandor GG, Lamb RK, et al. Pulmonary artery banding is not contraindicated in double inlet left ventricle with transposition and aortic arch obstruction. *Eur J Cardiothorac Surg*. 1995; 9:515-20.
7. Jensen RA Jr, Williams RG, Laks H, Drinkwater D, Kaplan S. Usefulness of banding of the pulmonary trunk with single ventricle physiology at risk for subaortic obstruction. *Am J Cardiol*. 1996;77:1089-93.
8. Odim JN, Laks H, Drinkwater DC Jr, George BL, Yun J, Salem M, et al. Staged surgical approach to neonates with aortic obstruction and single-ventricle physiology. *Ann Thorac Surg*. 1999;68:962-7; discussion 8.
9. Karl TR, Watterson KG, Sano S, Mee RB. Operations for subaortic stenosis in univentricular hearts. *Ann Thorac Surg*. 1991;52:420-7; discussion 7-8.
10. Lacour-Gayet F, Serraf A, Fermont L, Bruniaux J, Rey C, Touchot A, et al. Early palliation of univentricular hearts with subaortic stenosis and ventriculoarterial discordance: the arterial switch option. *J Thorac Cardiovasc Surg*. 1992;104: 1238-45.
11. Lee MG, Kowalski R, Galati JC, Cheung MM, Jones B, Koleff J, et al. Twenty-four-hour ambulatory blood pressure monitoring detects a high prevalence of hypertension late after coarctation repair in patients with hypoplastic arches. *J Thorac Cardiovasc Surg*. 2012;144:1110-6.
12. Rakhra SS, Lee M, Iyengar AJ, Wheaton GR, Grigg L, Konstantinov IE, et al. Poor outcomes after surgery for coarctation repair with hypoplastic arch warrants more extensive initial surgery and close long-term follow-up. *Interact Cardiovasc Thorac Surg*. 2013;16:31-6.
13. Jegatheeswaran A, McCrindle BW, Blackstone EH, Jacobs ML, Lofland GK, Austin EH III, et al. Persistent risk of subsequent procedures and mortality in patients after interrupted aortic arch repair: a Congenital Heart Surgeons' Society study. *J Thorac Cardiovasc Surg*. 2010;140:1059-75.e2.
14. de Leval MR, Deanfield JE. Four decades of Fontan palliation. *Nat Rev Cardiol*. 2010;7:520-7.
15. National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. *Pediatrics*. 2004;114(suppl 4th report):555-76.
16. Chobanian AV, Bakris GL, Black HR, Cushman WC, Green LA, Izzo JL Jr, et al. The Seventh Report of the Joint National Committee on Prevention, Detection,

Evaluation, and Treatment of High Blood Pressure: the JNC 7 report. *JAMA*. 2003;289:2560-72.
17. O'Sullivan JJ, Derrick G, Darnell R. Prevalence of hypertension in children after early repair of coarctation of the aorta: a cohort study using casual and 24 hour blood pressure measurement. *Heart*. 2002;88:163-6.
18. Jacobs ML, Rychik J, Murphy JD, Nicolson SC, Steven JM, Norwood WI. Results of Norwood's operation for lesions other than hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg*. 1995;110:1555-61; discussion 61-2.
19. Serraf A, Conte S, Lacour-Gayet F, Bruniaux J, Sousa-Uva M, Roussin R, et al. Systemic obstruction in univentricular hearts: surgical options for neonates. *Ann Thorac Surg*. 1995;60:970-6; discussion 6-7.
20. Tchervenkov CI, Tsang JC. Surgical treatment of single ventricle with aortic arch obstruction in early life. *Adv Cardiac Surg*. 1999;11:193-219.
21. Brawn WJ, Sethia B, Jagtap R, Stumper OF, Wright JG, De Giovanni JV, et al. Univentricular heart with systemic outflow obstruction: palliation by primary Damus procedure. *Ann Thorac Surg*. 1995;59:1441-7.
22. McElhinney DB, Reddy VM, Silverman NH, Hanley FL. Modified Damus-Kaye-Stansel procedure for single ventricle, subaortic stenosis, and arch obstruction in neonates and infants: midterm results and techniques for avoiding circulatory arrest. *J Thorac Cardiovasc Surg*. 1997;114:718-25; discussion 25-6.
23. d'Udekem Y, Xu MY, Galati JC, Lu S, Iyengar AJ, Konstantinov IE, et al. Predictors of survival after single-ventricle palliation: the impact of right ventricular dominance. *J Am Coll Cardiol*. 2012;59:1178-85.
24. Azakie T, Merklinger SL, McCrindle BW, Van Arsdell GS, Lee KJ, Benson LN, et al. Evolving strategies and improving outcomes of the modified norwood procedure: a 10-year single-institution experience. *Ann Thorac Surg*. 2001;72:1349-53.
25. McGuirk SP, Griselli M, Stumper OF, Rumball EM, Miller P, Dhillon R, et al. Staged surgical management of hypoplastic left heart syndrome: a single institution 12 year experience. *Heart*. 2006;92:364-70.
26. Mahle WT, Spray TL, Wernovsky G, Gaynor JW, Clark BJ III. Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. *Circulation*. 2000;102(Suppl 3):III136-41.
27. Heinle JS, Carberry KE, McKenzie ED, Liou A, Katigbak PA, Fraser CD Jr. Outcomes after the palliative arterial switch operation in neonates with single-ventricle anatomy. *Ann Thorac Surg*. 2013;95:212-8; discussion 8-9.
28. Amin Z, Backer CL, Duffy CE, Mavroudis C. Does banding the pulmonary artery affect pulmonary valve function after the Damus-Kaye-Stansel operation? *Ann Thorac Surg*. 1998;66:836-41.
29. Daenen W, Eyskens B, Meyns B, Gewillig M. Neonatal pulmonary artery banding does not compromise the short-term function of a Damus-Kaye-Stansel connection. *Eur J Cardiothorac Surg*. 2000;17:655-7.